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Techniques in Vascular and Interventional Radiology

Percutaneous Treatment of Lymphatic Malformations

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Lymphatic malformations are slow-flow vascular anomalies composed of dilated lymphatic channels and cysts of varying sizes. Percutaneous treatments, particularly sclerotherapy, play an important role in the treatment of these lesions, often obviating the need for surgical intervention. Owing to the complex nature of these lesions, a multidisciplinary approach should be used to guide diagnosis and management. This submission focuses on the workup and treatment of pediatric lymphatic malformations at our institution, with a focus on sclerotherapy. Therapeutic outcomes and the management of postprocedural complications are also discussed.

KEYWORDS Sclerotherapy, Lymphatic malformations, Pediatric

Background

Lymphatic malformations (LMs) are slow-flow vascular anomalies composed of dilated lymphatic channels and cysts. LMs do not demonstrate discrete communications with the normal lymphatic or venous systems. Depending on cyst size, they are classified as macrocystic, microcystic, or combined lesions (Fig. 1). Although there is no consensus definition or size cutoff of macrocystic vs microcystic LMs, the term macrocystic is generally reserved for lesions that are easily amenable to aspiration, which is usually 1-2 cm.[1,2] LMs are often transspatial in nature and can range in size from localized to diffusely infiltrating. Recent studies estimate the incidence to be 1/2000-4000 live births, without significant difference in sex.[3] Owing to the complexity of these lesions, our institution favors a multidisciplinary approach to clinical evaluation and treatment.

Clinical Evaluation

Presentation and Clinical Assessment

Clinical presentation depends primarily on the size and location of the malformation. Although LMs may involve any part of the body, most (48%-75%) are found in the cervicofacial region and 20%-42% are found in the extremities.[4,5] If large enough, lesions may be detected as early as the first trimester on prenatal ultrasound or prenatal magnetic resonance imaging (MRI). Those not identified prenatally would usually present shortly after birth as a soft tissue mass. The vast majority of the lesions present before the age of 2 years.[6]

Like other vascular malformations, LMs are congenital and grow proportionally with the child. If the lesions are small, patients may not present to the clinic until later in childhood when a lesion-related complication, such as infection or spontaneous hemorrhage, brings it to attention. Rarely, lesions would return to their prior size after resolution of the insult, going unnoticed until a repeat complication. LMs that are near joints may cause difficulty in ambulation, noticeable when the child is learning to walk or crawl.

As with other vascular malformations, management is best addressed in the setting of a vascular anomalies clinic, which is commonly staffed by interventional radiology and plastic surgery, with support from services such as dermatology, otorhinolaryngology, general surgery, ophthalmology, and oncology. On physical examination, LMs are solitary or multifocal soft masses with normal overlying skin. There are cases, however, where involvement of the dermal lymphatics results in the formation of dermal vesicles or angiokeratomas, which themselves may ooze blood-stained lymphatic fluid for a prolonged period or become superinfected (Fig. 2A). The skin may show a bluish discoloration if hemorrhage is present or show warmth in cases of infection

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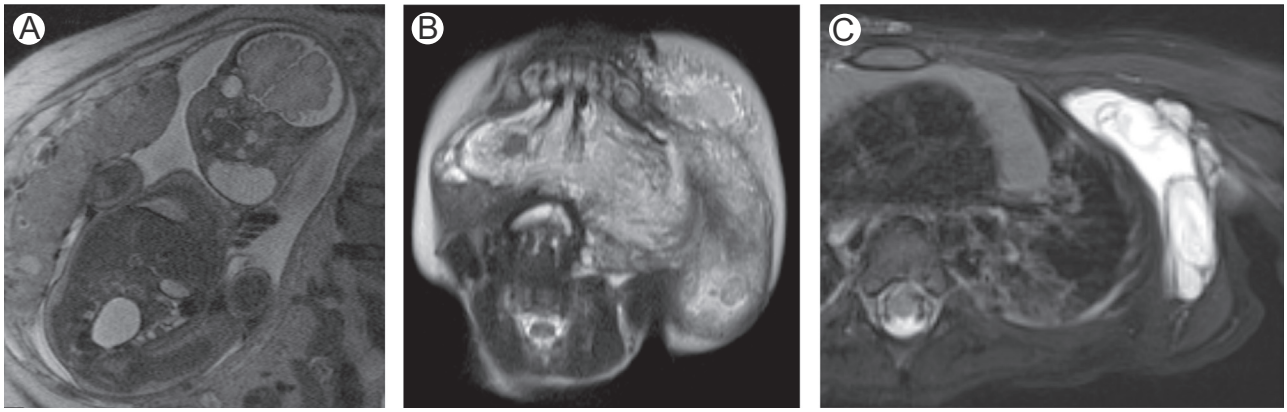

Figure 1 T2-weighted images showing different types of lymphatic malformations. (A) Prenatal MRI shows a macrocystic malformation involving neck. (B) A 3-month-old male with a cervical microcystic LM extending into the floor of the mouth. (C) A 10-month-old male with a left axillary mixed LM.

(Fig. 2B). LMs do not have a palpable pulse or thrill to suggest a high-flow component. Macrocystic LMs may transilluminate with a flashlight. Physical examination of the LM should primarily focus on identifying the location, extent and the relationship of the lesion to nearby structures. Pretreatment clinical photographs should also be obtained to document a baseline appearance of the lesion.

Imaging Assessment

Ultrasound is useful in the workup and to monitor treatment effect of LMs, particularly for smaller superficial lesions. Therefore, the baseline ultrasound should detail the extent, components and size of the lesion. The appearance of LMs on ultrasound depends on the cyst size. Macrocystic lesions appear as compressible, predominantly anechoic cysts with thin septations. Debris, which may result from hemorrhage or infection, may layer dependently within the cavities. Although blood vessels are detectible in cyst walls and septa, there should be no internal vascularity. In contrast, microcystic lesions consist of tiny cavities that produce a hyperechoic, solid appearance.

Most of our patients undergo an MRI study before treatment. MRI not only clarifies the diagnosis but also provides a better assessment of the extent of the lesion, therefore providing guidance for treatment. On MRI, LMs may transgress soft tissue planes and should follow fluid signal on all sequences in uncomplicated lesions. Fluid-protein levels may also be present. Hyperintensity on T1-weighted images may be seen in the setting of hemorrhage. The lesion should not contain flow voids, and there should be no filling defects to suggest the presence of a thrombus or phlebolith. There may be varying degrees of interstitial fatty elements as well as enhancement of septations. When LMs are predominantly microcystic they can have the appearance of a solid lesion and may mimic a tumor. A venous malformation can coexist with lymphatic components in a subset of patients, yielding the term "venolymphatic" malformation.

It is imperative to consider other differential diagnoses before initiating therapy. Mimickers of LMs in the young child include complex neoplasms such as teratomas, lipoblastomas, and rhabdomyosarcomas. These lesions are expected to show more solid and enhancing components. Nonneoplastic mimickers include cystic lesions such as dermoids or epidermoids and ciliated cysts.[7] Most of these lesions can be differentiated based on clinical history, location, and appropriate imaging. In difficult cases, percutaneous biopsy should be considered for definitive diagnosis.

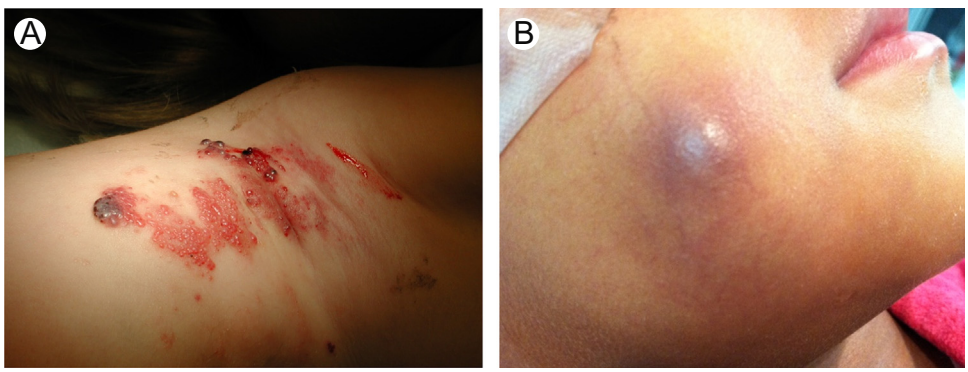

Figure 2 (A) Dermal vesicles (angiokeratomas) overlying a lymphatic malformation, which have become infected. (B) Bluish discoloration overlying a LM as a consequence of internal hemorrhage. (Color version of figure is available online.)